

Position Statement

AIS Support Group

8th May 2001

In Confidence

Re: Proposal to promote Hammersmith (O. Charlotte's & Chelsea Hospital) as the "National Centre for Adolescent and Adult Females with Congenital Abnormalities of the Genital Tract"

We are very concerned to hear of a proposal on the part of National Specialist Commissioning Advisory Group (NSCAG) at the UK Department of Health to promote Queen Charlotte's and Chelsea Hospital – QCCH (Hammersmith NHS Trust) as the "National Centre for Adolescent and Adult Females with Congenital Abnormalities of the Genital Tract", a move that would presumably compromise the well established role of the UCL/Middlesex and Leeds (General Infirmary and St. James's) Hospitals as multi-disciplinary centres of excellence in the treatment of intersex conditions. We set out below the reasons why we think the proposal is a seriously bad idea.

Introduction

The AIS Support Group (AISSG) is the only UK peer support group for a number of XY female intersex conditions such as androgen insensitivity syndrome (AIS), XY gonadal dysgenesis (Swyer's syndrome), 5-alpha reductase deficiency and leydig cell hypoplasia; and we also have members with XX female (non-intersex) conditions such as Mayer Rokitansky Kuster Hauser (MRKH) syndrome, Mullerian dysgenesis/aplasia and vaginal atresia.

We have enjoyed a strong and mutually beneficial working relationship with clinical teams at UCL/Middlesex and Leeds Hospitals since early 1995. At that time our group was still in the early stages of its development and was working hard to bring Androgen Insensitivity Syndrome (old name Testicular Feminization Syndrome) and similar conditions out into the open and make intersex less of a shameful and taboo subject. We have had no such productive relationship with QCCH.

Our Early Dealings with QCCH

Requesting Input for our Literature

In 1993, we wrote to a consultant gynaecologist at QCCH (and to a number of clinicians at other hospitals) explaining that we were formalizing our group (which had been started in a small, homespun way in 1988), were preparing a factsheet for parents and patients, and would welcome his input. We were particularly interested in obtaining his contribution because we had

heard on the grapevine that he was involved in some sort of support group for patients with problems like AIS and we were concerned that our efforts might duplicate or conflict with his.

Discouraging Feedback

We asked him about the nature of his group, expressing a wish not to 'tread on his turf' as it were, but were not able to elicit any response on this in spite of repeated follow-up communications. However we did receive an early reply discouraging us from our attempts to increase awareness of AIS. He said he was happy for us to form a support group for *parents* but he was clearly very unhappy about XY female patients getting to know their diagnosis. See "Psychological Paternalism" in ALIAS No. 3, Winter 1995. (ALIAS - *Looking At AIS*, is the support group's newsletter, see http://www.medhelp.org/www/ais/13_ALIASES.htm).

The QCCH Support Group

We eventually found out from other quarters that the support group run by QCCH was not actually a peer support group for AIS or intersex conditions (they would never use terms like this anyway, in view of their secrecy/paternalism regarding intersex diagnoses) but was a hospital-controlled facility whereby women with vaginal hypoplasia/agenesis¹ would meet once or twice a year, in the hospital, to discuss their progress with vaginal lengthening by pressure dilatation.

Paternalism vs Openness

One of our AIS group members who herself had vaginal hypoplasia tried to join the QCCH support group, purely for her own benefit in dealing with her own situation, but this was obstructed all along the way (her repeated enquiries were ignored). Having persisted, she was summoned for an 'interview' with the cons. gynaecologist (who presumably knew she was an AISSG member) at which she was grilled as to her motives and told that she could only attend the meetings if she promised not to reveal diagnostic information to the others in the meetings (something she had no intention of doing anyway) since most of them had not been told their diagnosis. She left his office in tears.

Since many of QCCH's XY female patients have not, we believe, been told their diagnosis, their "support group" cannot facilitate true peer support, in the holistic sense of addressing all aspects of the patient's condition with an honesty that will allow her to make sense of her situation, assign it a label, gain the comfort of in-depth discussion with others..... probably grieve (a natural and cathartic process).....and thus move on with her life.

The BBC "Dark Secrets" Documentary

In 1995 we heard that the QCCH consultant had declined an invitation from the BBC to take part in a documentary called "XY Women" (part of a series called "Dark Secrets") and, moreover, had actively tried to discourage the BBC from making the film. The programme was made, with the help of other clinicians (e.g. Prof. Howard Jacobs, then Prof. of Reproductive Endocrinology at the Middlesex Hospital, now retired) and was broadcast on BBC2 on 29th Aug 1996 (see "BBC Documentary on AIS" in ALIAS No. 6, Winter 1996).

The BBC provided a helpline and passed enquiries on to us. This film has proved to be an important vehicle by which many AIS women have for the first time been able to understand

1. short/absent vagina: one of the features of AIS and some related conditions.

their condition, introduce it to family and friends, and find peer support by locating our support group. It is very frequently mentioned as a 'life-saver' by people making contact with us.

Fear of the Truth

We began to understand that the cons. gynaecologist had inherited from his predecessor, Prof. Dewhurst, the paternalistic philosophy of withholding the truth from XY female patients about their diagnosis. He feared, one assumes, a flood of angry patients complaining about this, if AIS and other intersex conditions were to come out from behind the curtain of secrecy and half-truths with which he (and other clinicians) had shrouded them hitherto.

Our Collaboration with UCL/Middx and Leeds

First Contact with UCL/Middx Clinicians

Members of our group were invited by Dr Richard Stanhope (paed. endocrinologist, Gt. Ormond Street Children's Hospital) to take part in a conference on "*The Management of Intersex into Adult Life - especially androgen insensitivity/testicular feminization*" at the Royal Society of Medicine London on 26th April 1995 (see "Report on RSM Symposium" in ALIAS No. 2, Summer 1995).

This was possibly the first occasion on which a patient support group had been invited to participate in a medical conference on equal terms with clinicians. We heard afterwards that some of the more 'old-school' members of the audience had not approved of this but we received highly appreciative letters from a few clinicians including Dr Gerard Conway of UCL/Middlesex and Mr Adam Balen of Leeds praising our presentation and saying how good it was that we'd been involved (see "Doctors Write to Us" in ALIAS No. 3, Winter 1995). The QCCH consultant was a speaker at the conference. We received no such message from him. This was the start of our working relationship with the UCL/Middx and Leeds clinical teams.

Gave up Contact with QCCH

We quickly gave up trying to liaise with QCCH and concentrated all our efforts on taking up the interest shown in our endeavours by the clinicians at UCL/Middlesex and Leeds. From the start, UCL/Middx and Leeds have operated a policy of openness and truthful disclosure that is vital in enabling patients to "put the final pieces in the jigsaw, start closing the box and get on with their lives" and has been very much appreciated by patients.

Dr Gerard Conway introduced us to his colleagues Miss Sarah Creighton (cons. gynaecologist), Miss Melanie Davies (cons. gynaecologist) and Dr Lih-Mei Liao (clin. psychologist); and contact was established with Ms Julie Alderson¹ (clin. psychologist) who works with Mr Adam Balen (cons. gynaecologist) in Leeds. More recently we have also met UCL/Middx clinicians Dr Peter Hindmarsh (paed. endocrinologist) and Dr Caroline Brain (paed. endocrinologist) and we have remained in touch with Dr Richard Stanhope. Since mid-1997 we have been actively involved with these clinicians in the following initiatives:

Multi-Disciplinary Patient Care

The first meeting to discuss the development of multi-disciplinary clinics for XY female patients took place on 6th Aug 1997 between Gerard Conway, Sarah Creighton, Melanie Davies, Lih-Mei Liao, seven representatives from AISSG and two from the support group for

1. Now Dr Julie Alderson

Congenital Adrenal Hyperplasia (see "Middlesex Hospital Forum" in ALIAS No. 10, Winter 1997).

A second meeting took place on 20th Jan 1999 (see "Middlesex Hospital Update" in ALIAS No. 14, Spring 1999) by which time Dr Cathy Minto had joined the UCL/Middx team as gynaecology research fellow and introduced the idea of doing a research study with the help of the support group.

The multi-disciplinary clinic (endocrinology, gynaecology, psychology) has now been successfully operating at UCL/Middx for a number of years. We feel that a multi-disciplinary approach is long overdue in the treatment of intersex conditions and that this must take place in a centre that has expertise not only in gynaecology, but in endocrinology (paediatric and adult) and in clinical psychology. The psycho-social ramifications on intersex are large and have only recently started to be addressed by clinicians. The clinicians at the UCL/Middx and Leeds centres are at the forefront of this shift in emphasis.

The psychological management of conditions like AIS amounts to rather more than repeatedly telling such patients they are "just normal women". Yes, in many senses they are, but this realisation comes about by meeting others similarly affected, *with all the cards on the table*, and not by such controlling and paternalistic exhortations which, paradoxically, only sow the seeds of doubt in the mind of the patients ("Methinks he doth protest too much").

Guest Speakers at our Group Meetings

Following the attendance of Dr Conway and Dr Stanhope at our 5th parent/patient group meeting in March 1997, the various UCL/Middx and Leeds clinicians have been regular guest speakers at our parent/patient group meetings, giving up their Saturdays to be with us.

AIS Patient's Charter

In 1997 Dr Stanhope drew up an AIS Patient's Charter for us (see copy on website and also "Draft AIS Patient's Charter" in ALIAS No. 10, Winter 1997).

AISSG Recommends UCL/Middx/Leeds

In 1998 we started developing our web site at <http://www.medhelp.org/www/ais> and from an early stage listed our clinical colleagues on our Recommended Clinicians page. We have had no further contact at all with QCCH since the early/mid 1990's so we do not recommend any clinicians from there.

Research Studies

The idea of the support group helping with research studies was first suggested by Gerard Conway and Lih-Mei Liao following our 7th parent/patient meeting in March 1998 (see "Research Plans" in ALIAS No. 12, Summer 1998).

We have now helped with the following research projects (see also Research Studies page on our web site):

Julie Alderson was a guest at our Sept 1998 group meeting where she discussed a proposed research study (see "Clinical Psychology Study" in ALIAS No. 14, Spring 1997). Completed in 2001, this study involved interviews with a number of group members and formed the basis of

her doctoral degree titled "*Women with Androgen Insensitivity Syndrome (AIS): A Qualitative Study*". Julie is now writing up the findings for journal articles.

Cathy Minto spoke of an intended research project in early 1999, as mentioned earlier. Her project started life as a study of vaginal hypoplasia but she found that there were so many additional areas of importance that its scope has widened since then. By March 2001 she had had 130 study questionnaires returned (about 50% being from members of support groups for AIS, Congenital Adrenal Hyperplasia and MRKH and 50% from clinic patients), she had examined a number of the women, and was starting to write up research papers and conference presentations based on the findings.

Sue Smith (clin. psychology doctorate candidate, University of East London) linked up with Cathy some months later and developed a parallel study on psychological aspects (see "Clinical Research Update" in ALIAS No. 15, Summer 1999. This ALIAS article contains updates on the studies of Lih-Mei and Cathy, and announces Sue's study). Sue's study was completed in early 2001 and used in her doctorate thesis which was titled "*Experiences of sexuality reported by women with intersex conditions who have undergone some form of genital modification: A tale of two sexes?*". Again, this study was based on interviews with AISSG members.

Gerard Conway first suggested a study of medical aspects of AIS, and involving a trial of Testosterone HRT in women with the complete form of AIS, in early 2000 (see "10th UK Meeting" in ALIAS No. 16, Spring 2000). By early 2001, he had completed a pilot study and was embarking on the main study.

The following ALIAS articles contain further updates on these various studies:

- "UK Research Studies" and "11th UK meeting" in ALIAS No. 17, Summer 2000
- "Research Studies Completed" in ALIAS No. 18, Spring 2001
- "Research Studies Progressing" in ALIAS No. 18, Spring 2001
- "12th UK Meeting" in ALIAS No. 18, Spring 2001

Polly Carmichael (paediatric clin. psychologist, Gt. Ormond Street) announced a research project to study how parents deal with information about AIS in their children (see "Parents' Research Study" in ALIAS No. 18, Spring 2001) Polly had attended our group meeting in March 1997 with Richard Stanhope and also our March 2000 meeting.

UCL/Middx Clinic 'Open Days'

We have taken part in occasional Middx/UCL clinic 'open days'. These are hospital-based discussion forums to which all the hospital's XY female patients are invited and at which the clinicians explain medical issues and AISSG representatives talk about the support group. Afterwards, the clinicians in various specialties are available for regular private consultations. See "Clinic of the Future?" in ALIAS No. 17, Summer 2000.

Upcoming AIS Symposium

In early 2000, the idea of holding an AIS Symposium (i.e. a full-scale medical conference) was suggested (see "AIS Symposium" in ALIAS No. 17, Summer 2000 and "UK AIS Symposium" in ALIAS No. 18, Spring 2001). By late 2000 plans were being made, jointly by UCL/Middx (Sarah Creighton) and an AISSG representative, for a conference titled "*Intersex in the New Millennium. A multi-disciplinary forum for all those involved*" to be held in early 2002. Invited specialists and AISSG will play an equal part and the conference will acknowledge the co-operative mode in which we have been working towards improved patient care.

Summary/Discussion

Fanning Out Expertise

The plan for improved patient services, that the UCL/Middx clinicians discussed with us in mid-1997, was that the Leeds and London teams would become northern and southern UK centres of expertise and that this knowledge and experience could then be fanned out to a few other regional centres. A patient in the north of England will not want to trek down to a single national centre in London on a regular basis for clinic appointments. There are 30 or so specialist regional paediatric endocrinology centres, so it seemed sensible to work towards something similar for adult care (but with fewer centres).

We believe that intersex treatment belongs within the category of Specialised Commissioning (regional centres, usually 8-12, to concentrate expertise) rather than NSCAG (very rare conditions, 1 or 2 centres and a handful of patients). Incidence figures that we have to hand are 1 in 2,500 for Turner Syndrome, 1 in 5,000 for all variants of CAH and 1 in 20,000 for AIS. This represents a large workload for one centre.

QCCH is Still Reluctant to Acknowledge AIS?

We have no idea whether the attitude to truth disclosure at QCCH has changed over recent years but we would imagine that they will have had to 'go with the flow' to some extent because there has been a huge move on the part of intersex patient support and advocacy groups, e.g. Intersex Society of North America (ISNA), to increase clinical and public awareness of the needs of intersex patients, and of their right to such knowledge about themselves that will permit autonomy, and enable them to make sense of their situation and locate others who are affected.

We were quite surprised to discover about 6 months ago that QCCH now had a web site (<http://www.femgenab.org.uk/>). We even more surprised to see that they actually included a link to our site, which seemed like a reluctant admission on their part that their position regarding secrecy has become untenable. However, their site concentrates on MRKH Syndrome (and CAH) and does not describe AIS or other intersex conditions at all. They also make a dubious statement to the effect that absence of the vagina is known as MRKH Syndrome. Could this be an attempt to pull the wool over the eyes of AIS patients with absent/short vaginas by making them think they have a less threatening XX condition like MRKH, rather than a dreaded XY condition?

Whilst the paternalistic attitude of QCCH towards intersex patients may have changed slightly over the last few years (and we're not even sure that it has – we have only ever had one enquirer, out of several hundred, who has said that QCCH referred her to us), we have no relationship or goodwill built up with them at all, and still regard them as an island of out-dated practice, marooned by a tide of increasing openness and equal partnership taking place elsewhere.

QCCH is not an Intersex Centre

We get the impression that QCCH has built a good and well-deserved reputation, started in Prof. Dewhurst's days, for treating XX women with MRKH Syndrome, Mullerian Dysgenesis etc. for their vaginal hypoplasia, and that whilst they obviously have *some* XY female (i.e. intersex) women patients, these are "painted with the same brush" when they in fact have significant additional problems that are not experienced by the former group. The teams at Leeds and at UCL/Middx are very much aware of the additional medical and psychological issues experienced by XY women compared to XX female patients with reproductive development

problems, many of the psychological problems ensuing from a denial on the part of some clinicians of the existence of intersex when talking with anyone apart from other doctors; a denial that gets passed on to parents, patients and society in general.

From what we know of the set-up at QCCH it has not earned the right to consider itself a centre for the treatment of intersex patients. We approve of their dedication to promoting the non-surgical pressure dilatation method, but vaginal agenesis/hypoplasia is only *one* aspect of *some* intersex conditions. What about specialist endocrinology services? Dr Conway's predecessor at UCL/Middx, Prof. Howard Jacobs, was very well respected as an expert in reproductive endocrinology who had a 'user-friendly' patient-centred approach, one that very much still operates in the UCL/Middx team today. A number of CAIS women are finding, for example, that they feel better on testosterone- than oestrogen-based HRT. The UCL/Middx clinicians respect this. Most other hospitals will probably not sanction this treatment, especially one that is stuck in a groove of denial of the XY/intersex nature of AIS when dealing with patients.

What about Informed Consent?

In conditions like AIS there is a slight risk of cancer in testes left in situ beyond adulthood but many clinicians have in the past used this as a 'excuse' for removing them prematurely, in infancy/childhood. The real reason is probably to avoid awkward explanations to an older patient. An increasing number of AIS patients are angry at having undergone gonadectomy earlier in life (but even as adults in some cases) without having been told their diagnosis and therefore without informed consent. Some are angry at having thus lost their source of endogenous testosterone. Some are considering litigation. They argue that if clinicians did this to a man there'd be an outcry and that every normal XX woman has a small risk of breast cancer, and yet breasts are not removed prophylactically. QCCH will find this an increasing problem if its paternalistic and secretive attitude continues.

Paediatric Endocrinology/Urology is Important

QCCH has a heritage as a women's (maternity) hospital. Although it will no doubt have paediatricians on hand to deal with problems in the neonatal period, we wouldn't think it has the same expertise in paediatric endocrinology and paediatric urology that is available at UCL/Middx via its collaboration with experts (e.g. at Gt. Ormond Street Children's Hospital) within the same NHS Trust. Experts such as Dr Stanhope (whose predecessor Dr David Grant was a world-renowned expert in intersex in children), Mr Christopher Woodhouse (cons. urologist), Mr Philip Ransley (cons. paediatric urologist) etc., all of whom work very closely with our other clinical colleagues.

Transition to Adolescent/Adult Care

An integration of paediatric with adolescent/adult services is very important in intersex conditions. The UCL/Middlesex was the first UK hospital to launch an adolescent medicine service, in early 1998. Sarah Creighton and Cathy Minto are active members of the new *British Society of Paediatric and Adolescent Gynaecology (BritSPAG)* started in early 1999. Sarah is a member of the Working Party set up in early 2001 by the *British Association of Paediatric Urology (BAPU)* to evaluate the practice of 'gender reinforcement' surgery in intersexed infants with so-called ambiguous genitalia (see "BAPU Working Party" in ALIAS No. 18, Spring 2001), and issue that is now hotly debated, largely as a result of pressure from surgically 'corrected' intersexed adults via support/advocacy groups.

Carrier Testing Facilities?

Dr Caroline Brain (recently joined UCL/Middx) used to work in Cambridge with Prof. Ieuan Hughes, an internationally known expert in the genetics of AIS. Prof. Hughes' research lab used to provide some level of carrier testing on an ad hoc basis but this is no longer feasible (and was always very long-winded and unsatisfactory since it was done as a favour on the back of research activities).

XX female relatives of AIS women thus experience difficulty obtaining hard information on their possible carrier status, and demand for this type of service is likely to grow. In the past, secrecy and denial on the part of clinicians and within families meant that XX relatives were not made aware of the genetic inheritance pattern of conditions like AIS, but this is changing. Dr Conway would like to set up an NHS carrier testing service at the UCL/Middx. He has access to the necessary expertise but not to the required funding. What plans does QCCH have to deal with this issue?

Conclusion

The relationship that has been built up between the UCL/Middx and Leeds clinicians and ourselves (and other support groups such as those for CAH) is an excellent model of patient/doctor co-operation that has already borne much fruit in only a few years. The general attitude *of...* and the multi-disciplinary service provided *by...* these teams is universally praised by the many group members who now attend the clinics there, and are the envy of group members overseas. You only have to read the many glowing testimonials included in our newsletter to realise how our group members' appreciation of these clinicians, and *their* appreciation of *our* efforts, can only indicate the wisdom of building on this foundation and of further *funding* and *expanding* the activities at these centres.

We cannot work with a single national centre that concentrates only on gynaecology, does not have expertise in the endocrinological aspects of a range of intersex conditions and is, as far as we know, still in the dark ages regarding the sea change in the psycho-social treatment of intersex conditions that has occurred in the last 5-10 years.

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